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THE TYPES OF PULMONARY STENOSIS AND THEIR CLINICAL RECOGNITION

Pulmonary stenosis has recently aroused a great deal of interest, because (a) it is a part of the tetralogy of Fallot, and the cyanosis associated with this syndrome was found to be correctible by surgical operation[1]; (b) it has been successfully attacked by direct valvulotomy[2]. Pulmonary stenosis is almost always a congenital malformation, although in some cases the stenosed valve has the appearance similar to acquired valvular disease.

The renewed interest in this subject led to a search for autopsied cases of pulmonary stenosis and to a reexamination of older cases of pulmonary stenosis reported in the literature[3, 4].

There are several varieties of pulmonary stenosis as it is seen at the autopsy table. The most common of them are:

(1) *Pulmonary hypoplasia.* Apparently due to an unequal division of the common arterial trunk, the pulmonary orifice and the pulmonary artery are smaller than normal. This is often associated with bicuspid pulmonary valves. The degree of pulmonary stenosis varies from a size slightly smaller than normal to moderate stenosis. Severe stenosis in this form is rare.

(2) *Simple valvular stenosis.* This form resembles rheumatic stenosis of other valves and is characterized by thickening of the valves and joining of their free margins near the commissures. It usually causes mild or moderate narrowing of the pulmonic ostium.

(3) *"Diaphragmatic" type of pulmonary stenosis.* This is the most interesting and characteristic form of pulmonary stenosis. It resembles simple valvular stenosis, but there is complete fusion of cusps forming a "dome-shaped," or a "cone-shaped" membrane, or diaphragm directed with its convexity towards the pulmonary artery. It is perforated by a central opening which varies from 1 to 5 mm. in diameter. Its free margins are often the site of small bland vegetations. The diaphragm is relatively thin and delicate in younger subjects, but is thick and rigid in older individuals.

This and the preceding type of pulmonary stenosis are morphologically most similar to acquired valvular stenosis, and are thought by some to be due to fetal endocarditis. Both of these types occur much more often as a sole congenital malformation than as part of a syndrome consisting of multiple malformations.

(4) *Subpulmonic stenosis.* Below the normal, or hypoplastic pulmonary orifice with bi- or tricuspid pulmonary valves there is a horizontal fibrous membrane with a central perforation.

(5) *"Conus separate chamber"* (Abbott). The outflow tract of the right ventricle is separated from the main cavity by muscular bands forming a small chamber, "infundibulum" (Keith), communicating with the right ventricle by a small, or very small opening. At the upper end of this chamber there is a normal or a hypoplastic pulmonary orifice.

The last two forms of narrowing of the pulmonary outflow tract are most frequently combined with other lesions, notably with the tetralogy of Fallot.

The physiological effects of all of these forms of narrowing of the pulmonary orifice are identical: increased intraventricular pressure leading to right ventricular hypertrophy, and later, failure. If there exists an intracardiac communication, they facilitate right-to-left blood shunt. In addition, they are a common site of bacterial infection. The difference between the various forms lies in their effect on the pulmonary artery: subvalvular stenosis (types 4 and 5) have little effect upon the size of the pulmonary artery, which is found to be normal in size or hypoplastic. On the other hand, valvular forms (types 2 and 3), if severe, almost always lead to dilatation of the pulmonary artery, and occasionally of its branches. In some cases this dilatation assumes aneurysmal proportions.

Clinical patterns associated with pulmonary stenosis

Pulmonary stenosis is most often seen in combination with other congenital malformations. The best known syndrome is the tetralogy of Fallot, which is not only the commonest, but one allowing a relatively normal growth and development and survival to adult age. The various forms of pulmonary stenosis are a common finding in other, lesser known syndromes, such as transposition of the great vessels, trilocular heart, bilocular heart, or the common atrioventricular canal. From the clinical standpoint the important forms of pulmonary stenosis are those in which survival beyond infancy is possible and is frequent enough to make the clinical diagnosis of practical importance. The following syndromes can be considered of clinical importance:

A. *Pulmonary stenosis with closed septa.* 23 autopsied cases of this syndrome were found in the literature, two in infants, and twenty-one in patients whose age at death varied from 12 to 48 years. Nine patients died at the age of 30 or more. The cause of death in this group of patients was: cardiac failure in 7; bacterial endocarditis in 8; pulmonary tuberculosis in 2; 6 died of other causes. In 16 cases valvular stenosis was present, in 5 subvalvular. Of the 10 cases in which description of the pulmonary artery was available, 6 had a dilated artery.

Clinical features: This syndrome belongs to the noncyanotic group. In 17 cases cyanosis was absent, and among those were cases with most severe stenosis and cardiac failure. A critical review of the six cases in which cyanosis was reported reveals that only in one of these cases was long-standing, severe cyanosis present. On theoretical grounds the cause of cyanosis in the absence of intracardiac shunts is not clear.

Physical findings were reported as loud systolic murmurs and thrills in the upper left sternal border area, although occasionally systolic murmurs were reported at the lower part of the precordium, in-

cluding the apex. In five cases early diastolic blowing murmurs were described. The intensity of the second sound was usually diminished at the left side of the sternum.

Electrocardiograms revealed as a rule the pattern of right ventricular hypertrophy, and tall P-waves were often found. In one case right bundle-branch block was reported.

Roentgenological appearance of the cardiac shadow was that of a slightly or moderately enlarged heart with a characteristic enlargement of the pulmonary artery, which bulged prominently into the lung fields. Pulmonary branches were also frequently enlarged, but beyond these the lung fields were more radiolucent than normal due to a diminished vascular pattern. Fluoroscopy revealed usually absent or diminished pulsations of the large pulmonary vascular shadows.

B. *Pulmonary stenosis with closed interventricular septum and persistent patency of the foramen ovale.* An analysis of 29 autopsied cases of this syndrome shows a definite clinical pattern. Two patients died in infancy and in the twenty-seven, the age at death varied from 4 to 58 years. Five patients survived 30 years or more. Six patients died of heart failure, two of bacterial endocarditis, five of anoxia associated with severe cyanosis, and seven of pulmonary tuberculosis. In 28 of the 29 cases valvular stenosis was present, most frequently of the diaphragmatic type. Of the 15 cases in which the pulmonary vessels were described, 12 showed dilatation of the pulmonary artery.

Clinical features. The most important feature of this syndrome is cyanosis. Patency of the foramen ovale provides an important channel for a veno-arterial blood shunt, and in the majority of the cases the foramen was larger than the stenosed pulmonary ostium. Severe cyanosis with polycythemia and clubbing was almost invariably present in the older age group. In a few cases cyanosis was present since birth, in most of them it developed in late childhood, adolescence, or early adult life and grew progressively worse.

The presence of cyanosis is the fundamental distinguishing feature between cases of pure pulmonary stenosis with closed and with open foramen ovale. Physical findings, electrocardiographic and roentgenologic features are identical for both syndromes.

C. *Tetralogy of Fallot.* A group of 28 autopsied cases was analyzed which was limited to patients who survived for more than 15 years. The oldest patient died at 60. Three patients died of heart failure, 7 of bacterial endocarditis, 5 of pulmonary tuberculosis and of anoxia. Vulvular stenosis was present in 8, hypoplastic pulmonary artery with two pulmonic cusps in 7 cases — subvalvular stenosis was present in 8 cases and separate-conus-chamber type in one. The pulmonary artery was described as dilated in 5 cases and was normal or hypoplastic in 13.

Clinical features: As expected, cyanosis was a constant feature of the tetralogy and was present in all but one case. In some cases cyanosis had been present since birth, but in the majority of them it appeared in childhood or adolescence and became progressively worse. In two cases the onset of cyanosis occurred in adult life.

Physical findings: Murmurs appear to be less characteristic in this syndrome than in pure pulmonary stenosis. The location of a systolic murmur and thrill was reported more often at the apex and the lower left sternal border than at the "pulmonic area." Early diastolic murmurs were occasionally recorded.

Electrocardiographic findings were similar to pure pulmonary stenosis, characterized by right ventricular hypertrophy, tall P-waves, or occasionally right bundle-branch block. On the other hand, roentgenologically the tetralogy presents a marked contrast with pure pulmonary stenosis: The right ventricle is enlarged, frequently elevating the apex of the heart, as the so-called "Coeur en sabot," but the pulmonary vessels are small resulting in a deep concavity of the mid-portion of the left cardiac border.

As already mentioned, these three forms of pulmonary stenosis permit survival to adult age. Their incidence and relative frequency cannot be judged adequately from case collections from the literature. It is believed, however, that although pulmonary stenosis with patent foramen ovale is considerably rarer than the tetralogy of Fallot, it is the second most common cause of chronic congenital cyanosis in adults[3].

Summary

The importance of various forms of pulmonary stenosis lies in the fact that valvular stenosis, especially when severe, leads to dilatation of the pulmonary artery, while in sub-valvular stenosis such dilatation is uncommon. This observation can be utilized in differential diagnosis, for severe valvular stenosis occurs most frequently alone, unassociated with other malformations. Such pure pulmonary stenosis occurs in two forms: With and without persistent patency of the foramen ovale. In the first case, cyanosis is present and may be severe. In the second case, it is absent. In the tetralogy of Fallot pulmonary stenosis is usually milder, is frequently subvalvular or even appears as a mere hypoplasia of the pulmonary orifice and artery. The differential features of the three syndromes associated with pulmonary stenosis are presented.

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William H. Carnes, M.D. (Baltimore, Md.)

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NOTICE

Members of the American Heart Association and others are requested to contribute material (slides, blocks, tissues, gross specimens, etc.) together with related records from cases for the Registry of Cardiovascular Pathology of the Armed Forces Institute of Pathology, Washington, D. C.

At present, collection of material will be concentrated in the following categories:

1. **Congenital cardiovascular anomalies**
2. **Sub-acute bacterial endocarditis**
3. **Tumors of the Heart and Blood Vessels**
4. **"Diseases of the Collagen System" as polyarteritis nodosa, temporal arteritis, disseminated lupus erythematosus, scleroderma and amyloid disease.**

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